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CASE REPORT

Pressure-Lowering Effect of Fistula Occlusion in a Patient with Secondary Glaucoma Due to an Intracranial Arteriovenous Fistula

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ABSTRACT

Introduction: This case report presents the pressure-lowering effect of transluminal fistula occlusion in a patient suffering from secondary glaucoma due to carotid cavernous fistula (CCF).

Case Report: A 76-year-old Caucasian woman presented with dilated epibulbar vessels with elevated intraocular pressure (IOP, >30 mmHg) as well as glaucomatous excavations of the optic disc in both eyes. Cerebral digital subtraction angiography revealed a CCF with bilateral orbital communication. Preoperative diurnal pressure profiling showed an average IOP of 25.8 mmHg (right eye) and 26.6 mmHg (left eye). Transluminal intervention and fistula occlusion led to a decrease in IOP of about 9 mmHg. A post-operative oculomotor nerve palsy regressed spontaneously.

Conclusion: Secondary glaucoma due to CCF might be affected by fistula occlusion. Therefore, it should be considered before any surgical glaucoma interventions are performed. Diurnal pressure profiling is an effective tool for monitoring therapeutic success.

Keywords: Carotid cavernous fistula; Diurnal pressure monitoring; Embolization therapy; Secondary glaucoma

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INTRODUCTION

Intraocular pressure (IOP) is regulated via a constant equilibrium of intraocular fluid production and efflux. The interaction between intra- and periocular vessels and their state of filling plays a major role in the pathogenesis of secondary open-angle glaucoma. Fluctuations in the pressure ratios of these vessels directly affect IOP [1, 2]. The

normal episcleral venous pressure is about 8 to 10 mmHg. Once the venous pressure has reached the level of the IOP, an isotonic increase in IOP develops with additional increases in venous pressure. Moreover, an increase of pulsatile pressure fluctuations can be observed. With congestion-caused secondary glaucoma, the intraocular fluid increases, including both the intravasal and extravasal volumes. The autoregulatory processes of pressure equalization become disrupted and are, therefore, difficult to target with drug therapy. To determine the cause, the following conditions are to be considered: congenital vascular malformations (e.g., Sturge–Weber syndrome), orbital or intracranial arteriovenous fistulas, idiopathic forms (so-called idiopathic elevated episcleral venous pressure or Radius–Maumenee syndrome) or thrombotic events within the perfusion area of the large venous vessels in the head and neck area (intracranially, e.g., cavernous sinus; extracranially, e.g., jugular vein, superior vena cava, or pulmonary vein) [3–5]. Aside from glaucomatous changes, there are frequently congested epibulbar vessels, chemosis, eyelid edema and orbital bruits. Gonioscopy typically shows an open chamber angle, frequently with a blood-filled Schlemm's canal. Drug therapy is mostly inefficient and surgical intervention is necessary. Complications such as choroidal effusion as well as expulsive bleeding arise frequently in this condition [3, 5].

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1964, as revised in 2013. Informed consent was obtained from the patient for being included in the study.

CASE REPORT

A 76-year-old female patient was referred to our clinic for consultation with eye reddening and glaucoma "resistant to drug therapy". Despite topical triple therapy, the IOP was above 30 mmHg on both sides. On an outpatient basis, an Nd:YAG laser iridotomy had been performed on both eyes, which showed no effect on IOP. In general, the patient suffered from arterial hypertension and atrial fibrillation. Initially, these general symptoms were not identified.

With a more focused interview, she reported tinnitus that persisted for the previous 6 years. The IOP was in need of treatment for 3 years. For the past year, the patient noticed an increasing reddening of both eyes. The patient could not recall past trauma. Connective tissue weakness had not been reported.

Ophthalmological examination showed a best corrected visual acuity of 1.0 (Snellen charts) on both sides. The IOP was 27 mmHg in both eyes. Slit lamp examination showed markedly dilated epibulbar vessels (Fig. 1a). Intraocular findings included bilateral iris hyperemia (Fig. 1b) as well as glaucomatous papillae (progressed on the right side, beginning on the left side; Fig. 1c). There were progressive glaucomatous vision field defects. The patient was admitted for in-patient glaucoma diagnostics (i.e., diurnal pressure profiling (DPP) including night measurements using Goldmann applanation tonometry), as well as a digital subtraction angiography (DSA). The DPP showed pressure values of 23 to 28 mmHg on the right side (average 25.8 mmHg) and 24 to 29 mmHg on the left side (average 26.6 mmHg). The DSA results showed a complex situation with two arteriovenous shunts within the area of the cavernous sinus, as well as an arteriovenous fistula between the

left external carotid artery and the jugular vein. The carotid cavernous fistula (CCF) corresponded to a Barrow type B (Fig. 2). Following an unsuccessful attempt at conservative therapy by manual compression treatment, a neuroradiological intervention was scheduled. In the meantime, the patient received systemic carboanhydrase inhibitors.

Transluminal embolization of the CCF was performed after failure of conservative therapy attempts. Post-operatively, a cavernous sinus thrombosis occurred, with paresis of the right-side oculomotor nerve (complete, inside and outside). Due to a hemodynamically relevant bleeding within the puncture area (femoral artery), an

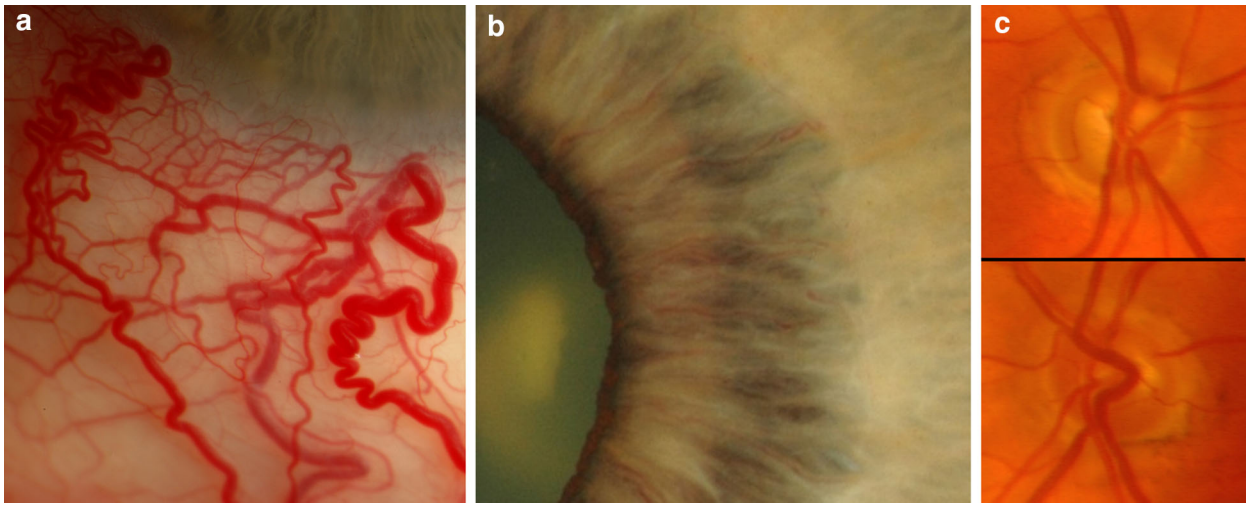


Fig. 1 Ophthalmological findings. **a** Dilated epibulbar vessels. **b** Congested vessels of the iris. **c** Fundus photograph of the papillae (*top* right eye, *bottom* left eye)

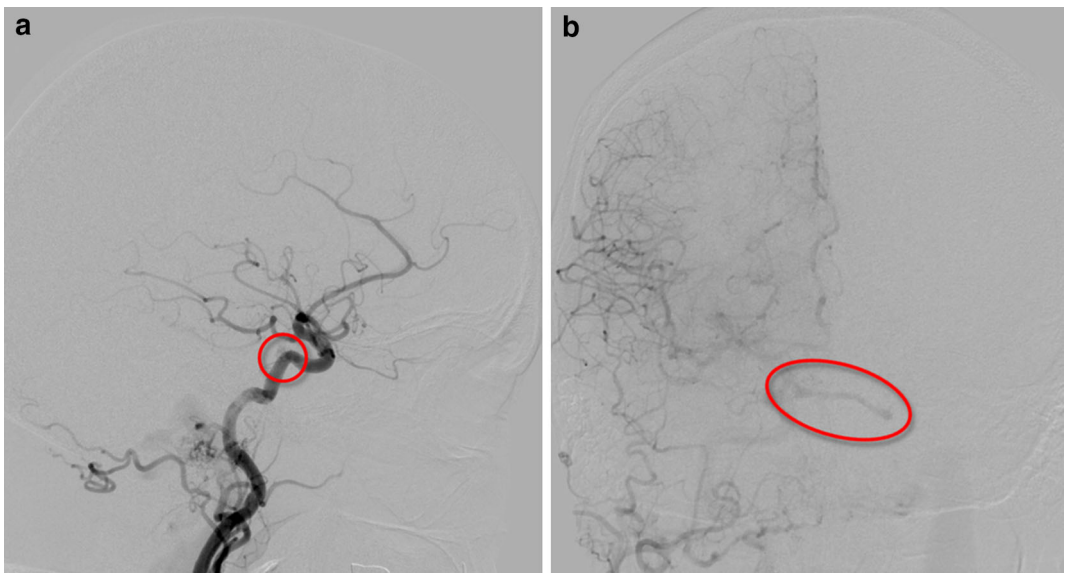


Fig. 2 Radiological findings (DSA). **a** Lateral view (fistula location labeled). **b** A–P view (retrograde filling of the anterior intercavernous sinus labeled). *DSA* digital subtraction angiography

intervention performed by the vascular surgery department became necessary.

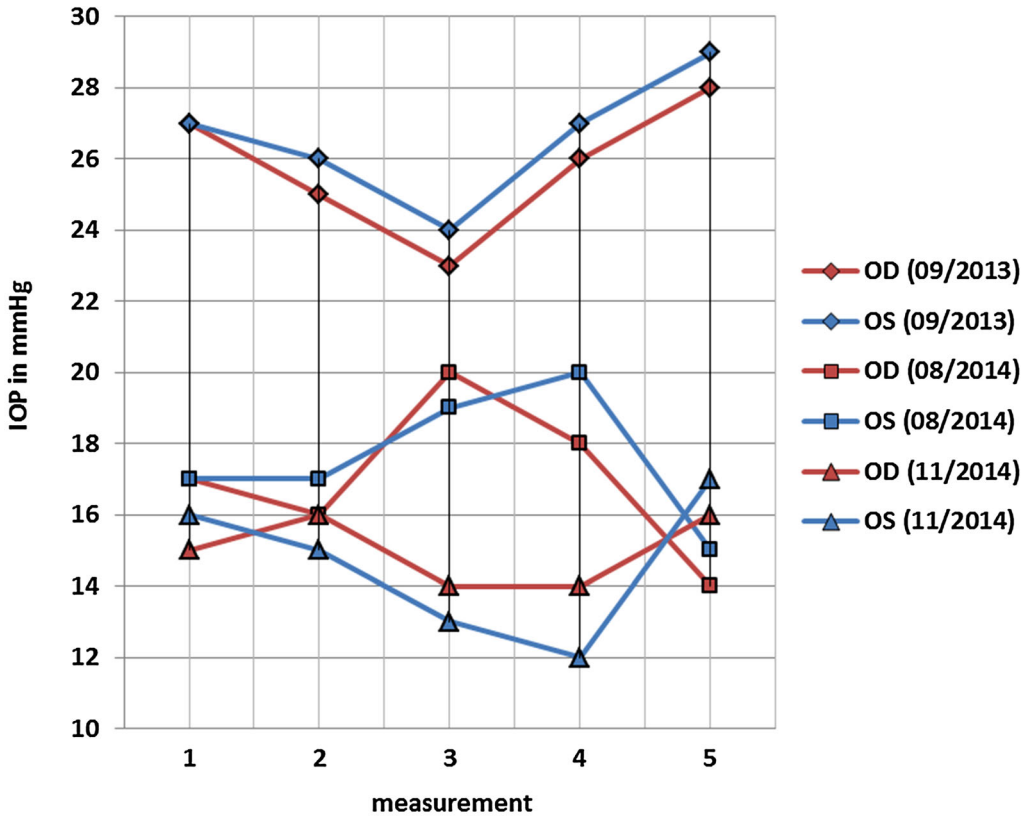
Ptosis and double vision showed rapid improvement. After 4 weeks, the patient was free of double vision. A discrete ptosis and an anisocoria remained. Six months after surgery of the CCF, a DPP was performed again. Although topical therapy only consisted of two anti-glaucomatous drugs, the IOP was the following: on the right side, 14 to 20 mmHg (average 17.0 mmHg); on the left side, 15 to 20 mmHg (average 17.6 mmHg). Thus, the IOP was lowered on the right side by a mean of 8.8 mmHg, and on the left side, by a mean of 9.0 mmHg. Three months later the DPP showed a mean pressure of 15.0 mmHg on the right side and 14.6 mmHg on the left side (Fig. 3). Threshold perimetry and measurement of the retinal nerve fiber layer did not indicate a disease progression.

DISCUSSION

CCF produces an intracranial arteriovenous shunt [6, 7]. Typical clinical signs and the emergence of secondary glaucoma should lead an ophthalmologist to suspect CCF. An imaging procedure (i.e., DSA) serves to ascertain the diagnosis. CCF can be classified according to cause (traumatic vs. spontaneous), hemodynamics (high flow vs. low flow) and anatomy (direct vs. indirect). Clinically, classification oriented toward anatomy (i.e., Barrow type A–D) has been established [6–8].

The cavernous sinus is a duplicature of the dura mater. It contains numerous pathways of conduction (i.e., cranial nerves III, IV, V_1, V_2 and VI, as well as the internal carotid artery). The venous connection to the orbita is provided by the superior ophthalmic vein. In CCF, there is more or less pronounced arterialization of this vessel, with corresponding reversal of flow and congestion [1, 3, 4, 9]. The consequence is an increase in episcleral venous pressure. This is transferred from the aqueous veins to the collecting vessels and Schlemm's canal and, thus, leads to an increase in IOP. Typical clinical signs may occur; i.e., red eye, pulsating exophthalmos and secondary glaucoma [7]. However, depending upon hemodynamics, symptoms may vary widely. High-flow (direct) fistulas usually occur acutely in connection with a trauma or rupture of an intracavernous aneurysm of the internal carotid artery. Most noticeable is a solid chemosis and an exophthalmos. Due to damage to the vasa nervorum, pareses may occur [6]. Dependent on the findings (e.g., impairment of visual acuity, IOP increase >40 mmHg, and cortical drainage path), there may be an emergency indication for fistula closure [9]. A low-flow (indirect) fistula results from a remodeling after cavernous sinus thrombosis with slowly developing arteriovenous shunts between branches of the internal carotid artery and the cavernous sinus. Usually older patients are affected. Pulsatile exophthalmos does not occur. Symptoms develop subtly, in which approximately one-third of the cases develop secondary glaucoma [3, 6, 9, 10]. Nevertheless, not every fistula requires treatment. However, regular ophthalmological monitoring is mandatory, as secondary glaucoma and vision impairments may develop subtly [3].

A retrospective multicenter investigation reports 14 eyes of 43 patients with CCF showing ocular manifestations [10]. Patients were recruited over 16 years. Of these about two-thirds developed secondary glaucoma. The most common symptom was conjunctival hyperemia (92.9%). Surgical fistula occlusion was done in only 4 of 9 cases. Here, three eyes had a good outcome of IOP control. If CCF



frame	side; period	t_1	t_2	t_3	t_4	t_5	IOP_{mean}	ΔIOP
1	OD (09/2013)	27	25	23	26	28	25,8	
	OS (09/2013)	27	26	24	27	29	26,6	
2	OD (08/2014)	17	16	20	18	14	17,0	8,8
	OS (08/2014)	17	17	19	20	15	17,6	9,0
3	OD (11/2014)	15	16	14	14	16	15,0	2,0
	OS (11/2014)	16	15	13	12	17	14,6	3,0

Fig. 3 IOP profiles compared before and after closure of the fistula. Time between measurements was 3 h. *DPP* diurnal pressure profile, *IOP* intraocular pressure, *OD* right eye, *OS* left eye, *t* time of measurement

closed spontaneously it was also predictive for good IOP outcome. Generally, anti-glaucoma treatments were more effective in eyes with CCF closure [10]. Therefore, these results may also focus on fistula closure as the primary therapy.

Currently, neuroradiological transluminal intervention is the therapy of choice. Over 90% of the fistulas can thereby be treated successfully [9]. Among the availabilities are fluid embolisates, coils or detachable balloons [9–12]. Arterial or venous access is possible and interdisciplinary approaches are to be favored [12, 13]. Aside from a very low immediate risks (e.g., stroke), thrombosis of the cavernous sinus may occur after intervention, as in the reported case. To reduce the risk for such complications, temporary therapeutic heparinization is indicated [14].

Patient support requires an interdisciplinary concept. For indication and evaluation,

standardized examinations are necessary. The clinical symptoms should be documented as well if possible (e.g., photographic documentation). For glaucoma diagnostics and post-operative evaluation, 24-h-intraocular pressure measurement is appropriate [15, 16]. Ideally and for improved reliability, this should be performed over 48 h or longer, in particular if vascular factors are pathogenetically involved [16, 17].

CONCLUSION

Intracranial arteriovenous shunts are not a homogeneous clinical entity and may manifest themselves in various forms. An intractable secondary glaucoma is a frequent complication and represents an indication for the treatment of CCF. Thereby, a clear pressure-lowering effect can be obtained. Fistula closure should thus take place before any surgical glaucoma interventions.

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Conflict of interest. Jens Heichel, Thomas Hammer, Laszlo Solymosi, Silvio Brandt and Iris Winter declare that they have no conflict of interest.

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